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Sinus of Valsalva Aneurysm Complicating Bacterial Endocarditis in an Infant: Diagnosis With Two-Dimensional and Doppler Echocardiography

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Sinus of Valsalva aneurysm is a rare complication of bacterial endocarditis in infancy and childhood. This report describes an infant with congenital aortic stenosis who developed bacterial endocarditis after abdominal surgery and placement of indwelling central venous catheters for long-term parenteral nutrition. Bacterial endocarditis in this infant was complicated by the development of an aneurysm of the left sinus of Valsalva. Surgical intervention was necessary because of gradual expansion of the aneurysm with compression of the adjacent right pulmonary artery and descending aorta. Two-dimensional and Doppler echocardiography proved useful for the initial diagnosis and serial follow-up of this unusual disorder and for its successful surgical management.

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Bacterial endocarditis is a rare entity in infants (1). This report describes an infant with congenital aortic stenosis who developed bacterial endocarditis with resultant aortic insufficiency and formation of a sinus of Valsalva aneurysm. Such an aneurysm is an unusual complication of bacterial endocarditis in adults and has not been previously reported in infants. Two-dimensional and Doppler echocardiography were useful in its initial diagnosis and subsequent management. Serial echocardiographic studies allowed close, noninvasive observation of the expanding lesion. Cardiac catheterization, a potentially dangerous procedure in a sick infant, was avoided.

Case Report

History. The patient was a 2.9 kg female infant born at 38 weeks' gestation to a 38 year old gravida 5, para 5 mother. The pregnancy, labor and delivery were uncomplicated. Apgar scores were 6 at 1 minute and 9 at 5 minutes. At birth, the infant was found to have ileal atresia, which was repaired at 1 day of age. Postoperatively, she required indwelling central venous catheters for parenteral nutrition. Recovery was uneventful. A heart murmur, thought to represent a ventricular septal defect and aortic stenosis (clinical and echocardiographic diagnosis), was also noted at birth. The infant was discharged from the hospital at 1 month of age and was well until 7 weeks of age when she developed symptoms of a mild upper respiratory infection (nasal stuffiness, tachypnea, axillary temperature of 38.5°C). She was initially treated with ampicillin but, when emesis developed, the treatment drug was changed to erythromycin. Over the ensuing week she remained tachypneic and irritable. A chest radiograph revealed cardiomegaly, and she was referred to our hospital for cardiologic evaluation.

Physical examination. On admission, the infant was acyanotic, pale and in mild distress. Her weight was 4.0 kg, height 54.5 cm, head circumference 37.5 cm. The heart rate was 160 beats/min, respiratory rate 60 to 70 breaths/min, blood pressure in the arms and legs 90 mm Hg systolic. The precordium was quiet and without thrill or abnormal impulse. The peripheral pulses were full in all limbs but not bounding. The first and second heart sounds were normal, and no click was heard. A grade 3/6 harsh systolic ejection murmur, which peaked in early systole and ended in midsystole, was heard in the second right intercostal space. A grade 2/4 high-pitched early diastolic murmur was heard in the second left intercostal space. The liver edge was palpable

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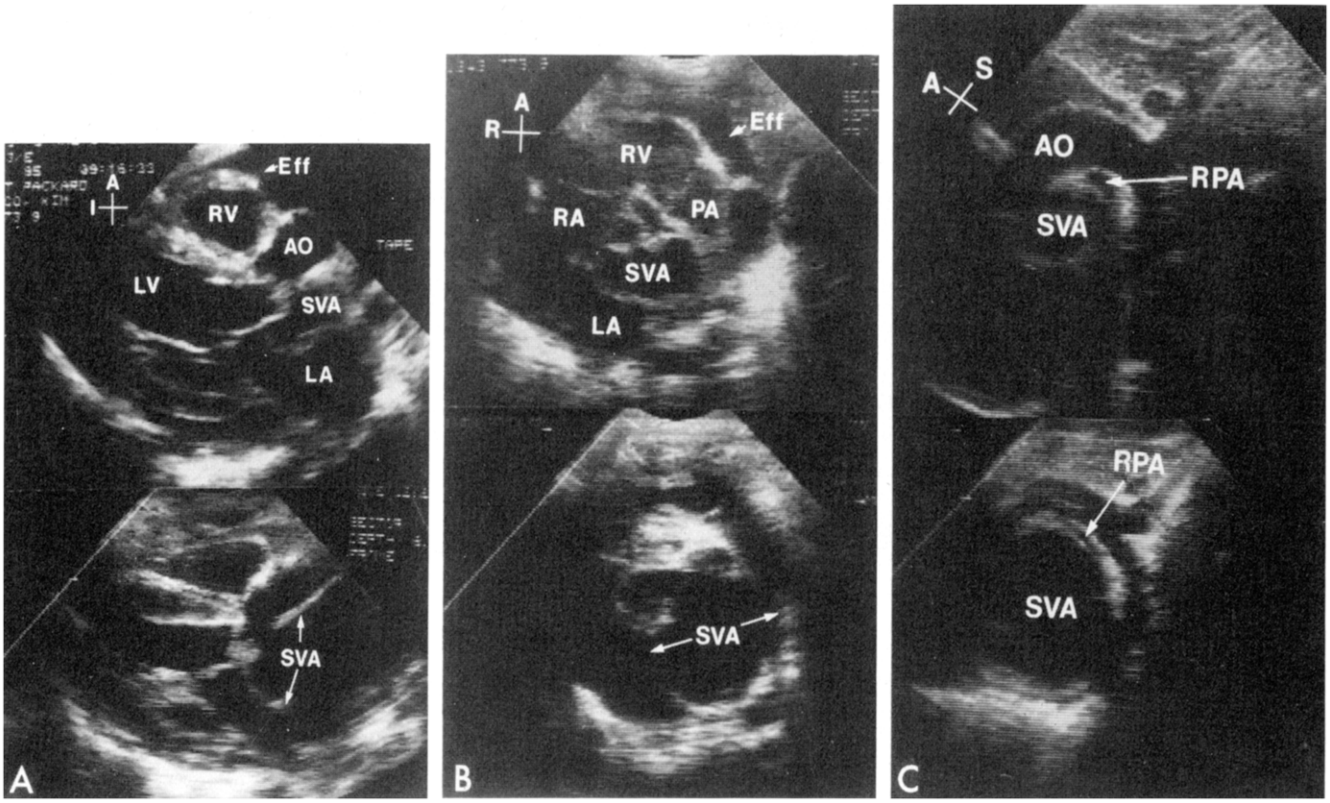

Figure 1. Serial two-dimensional echocardiograms. Parasternal long-axis views. **A: Top,** First hospital day. The dilation in the posterior portion of the ascending aorta (AO) is a sinus of Valsalva (SVA) aneurysm. There is a moderate-sized pericardial effusion (Eff), and the aortic valve leaflets are thickened. **Bottom,** 11th hospital day. The sinus of Valsalva aneurysm (SVA) has enlarged considerably. The aortic valve leaflets are prolapsing into the left ventricle (LV). **B:** Parasternal short-axis views. **Top,** First hospital day. The posterior and leftward dilation in the ascending aorta is a sinus of Valsalva (SVA) aneurysm. A moderate-sized pericardial effusion (Eff) is present. The aortic valve is bicuspid. **Bottom,** 18th hospital day. The sinus of Valsalva aneurysm has enlarged considerably and is impinging on the right pulmonary artery. **C:** Suprasternal long-axis views. **Top,** First hospital day. The sinus of Valsalva (SVA) aneurysm is seen bulging from the ascending aorta (AO) in a posterior and superior direction. There is some encroachment on the right pulmonary artery (RPA). **Bottom,** 18th hospital day. The right pulmonary artery (RPA) is markedly narrowed by the expanding sinus of Valsalva aneurysm. There is now encroachment on the descending aorta as well. A = anterior; I = inferior; LA = left atrium; P = posterior, PA = main pulmonary artery, R = right, RA = right atrium, RV = right ventricle; S = superior.

5 cm below the right costal margin, and the spleen was palpable 2 cm below the left costal margin.

Diagnostic studies. The chest radiograph showed marked cardiomegaly with normal pulmonary vascular markings. The electrocardiogram showed a QRS axis of +90°, no selective hypertrophy and inverted and flattened T waves throughout the chest leads. Laboratory studies revealed the following values: hemoglobin 6.6 g/100 ml, hematocrit 20.5%, white blood cell count 20,800 mm^3 with a normal differential and erythrocyte sedimentation rate 12 mm in 1 hour. A urinalysis was negative for red blood cells.

The two-dimensional echocardiogram showed concentric left ventricular hypertrophy and a thickened, domed, bicuspid aortic valve. An aneurysmal dilation was seen in the region of the left sinus of Valsalva (Fig. 1A and B). The right pulmonary artery was displaced superiorly and compressed by the aneurysm (Fig. 1C). A moderate pericardial effusion was present. The Doppler ultrasound examination showed a 4.2 m/s jet in the ascending aorta (predicted peak instantaneous aortic valve gradient 70 mm Hg) and evidence of mild aortic insufficiency (Fig. 2). The clinical and echocardiographic findings suggested severe aortic stenosis, mild aortic insufficiency and an aneurysm in the area of the left sinus of Valsalva.

Hospital course. The patient was presumed to have bacterial endocarditis of the aortic valve. She was initially treated with ampicillin, nafcillin and gentamycin. Six blood cultures grew *Staphylococcus epidermidis,* and the antibiotic drug was changed to vancomycin. Blood cultures were sterile 48 hours after initiation of therapy. Throughout her hospital course, she remained afebrile and asymptomatic. She was evaluated closely with serial echocardiograms. The sinus of Valsalva aneurysm was enlarging, gradually causing compression of adjacent structures (Fig. 1). The Doppler estimate of the pressure gradient across the aortic valve

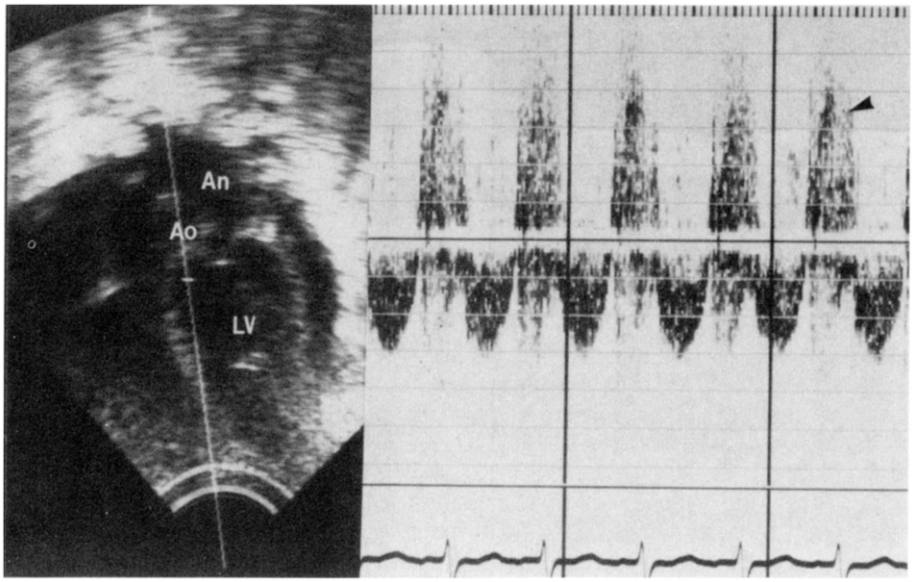

Figure 2. Pulsed Doppler examination of the left ventricular (LV) outflow tract on the first hospital day. The freeze-frame image (**left**) shows the position of the sample volume just beneath the aortic (AO) valve at the time of the recording. The spectral recording (**right**) shows normal forward flow out the left ventricle (LV) in systole below the baseline. In diastole, turbulent flow signals (**arrow**) are seen above the baseline, indicating flow toward the transducer from the aorta to the left ventricle. These flow signals represent aortic insufficiency. An = aneurysm. Transducer frequency = 3 MHz; calibration lines = 1.0 kHz.

remained unchanged throughout the first 2 weeks of hospitalization. Subsequently, the two-dimensional echocardiogram showed severe and progressive degenerative changes in the aortic valve. With the development of severe aortic valve prolapse and insufficiency, the left ventricular outflow tract gradient decreased to 30 mm Hg on Doppler examination (Fig. 3A). The aneurysm continued to expand, further compressed the right pulmonary artery and began to compress the descending aorta (Fig 1C). A 2.5 m/s jet (25 mm Hg predicted pressure gradient) was detectable on Doppler examination of the descending aorta by hospital day 18 (Fig. 3B). In addition, Doppler examination of the descending aorta showed continuous flow signals in systole and diastole indicating the existence of a pressure difference across the obstruction throughout the cardiac cycle.

Surgery. Because of echocardiographic evidence of an enlarging aneurysm with compression of adjacent structures despite adequate antimicrobial therapy, and because of fear

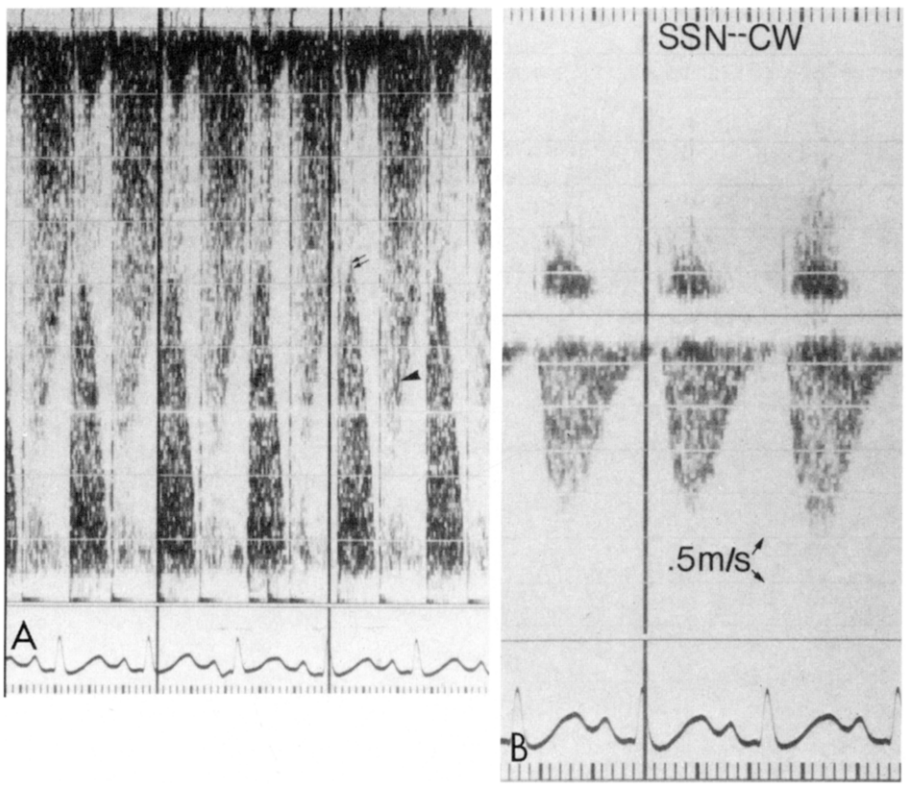

Figure 3. Continuous wave (CW) Doppler recordings obtained from the suprasternal notch (SSN). **A,** Ascending aorta, 18th hospital day. In systole, a jet (**small arrows**) is recorded above the baseline indicating flow toward the transducer from the left ventricle to the ascending aorta. The peak velocity of the jet is 2.7 m/s, indicating a 30 mm Hg pressure gradient across the aortic valve. In diastole, a high velocity jet (**large arrow**) is seen below the baseline, indicating regurgitant flow from the aorta to the left ventricle. As assessed by the pulsed Doppler system and a mapping technique, the amount of aortic insufficiency had increased dramatically since the initial studies. The Doppler recording is displayed with a zero shift baseline and calibration lines at 0.5 m/s intervals. **B,** Descending aorta, 18th hospital day. A 2.5 m/s jet (25 mm Hg predicted pressure gradient) is recorded in the descending aorta. The continuous flow signals throughout systole and diastole are characteristic of an aortic obstruction.

that the aneurysm would rupture, surgery was performed 4 weeks after admission. Cardiac catheterization and angiography were considered an additional risk in this infant and were not deemed necessary because of the clarity of the echocardiographic images. With the patient on cardiopulmonary bypass, a large aneurysm of the left sinus of Valsalva was found. The aneurysm displaced the right pulmonary artery superiorly and posteriorly. The aortic valve was bicuspid, and the posterior leaflet was destroyed; however, the anterior leaflet was intact. The aneurysm was dissected and resected. The native aortic valve was sutured closed and a valved conduit (Carpentier-Edwards 12 mm) was placed from the left ventricular apex to the ascending aorta. The infant's postoperative course was uncomplicated, and she went home 19 days after surgery. At the time of hospital discharge, the two-dimensional echocardiogram showed a hypertrophied left ventricle with normal systolic function. There was no insufficiency or stenosis of the valved conduit. Blood flow velocity in the descending aorta was normal (1.3 m/s).

Discussion

Bacterial endocarditis is unusual in infants (1) and is associated with a high mortality rate (2). It occurs more commonly in infants with congenital heart disease (3) and has been associated with long-term indwelling central venous catheters (4). Our patient had the unusual complication of a sinus of Valsalva aneurysm. This complication of bacterial endocarditis occurs uncommonly in adult patients (5) but has not been associated with bacterial endocarditis in infants.

Differential diagnosis. The echocardiogram showed an aneurysmal dilation in the area of the left sinus of Valsalva. The differential diagnosis of an aneurysmal dilation in the area of the aortic root includes aortico-left ventricular tunnel and aortic root dissection. In aortico-left ventricular tunnel, the right sinus of Valsalva is usually involved, and the tunnel between the aorta and left ventricle can usually be visualized on the two-dimensional echocardiogram as a channel or opening separate from the aortic valve (6,7). In our patient, the area of aneurysmal dilation involved the left sinus of Valsalva and communicated with the aortic valve. In aortic root dissection, an intimal flap can usually be seen on the two-dimensional echocardiogram. An intimal flap was not seen on the echocardiogram of our patient.

Management. The two-dimensional and Doppler echocardiograms allowed us to follow this patient serially and noninvasively, thus avoiding cardiac catheterization during a high risk period. The echocardiographic examinations accurately revealed progressive dilation of the sinus of Valsalva aneurysm despite adequate antimicrobial therapy. The Doppler examination showed evidence of progressive, hemodynamically significant compression of the adjacent right pulmonary artery and descending aorta. Surgical intervention became necessary although there had not been adequate time for complete healing of the aorta. At surgery, a large portion of the aortic valve and the left wall of the ascending aorta were composed of friable, necrotic tissue. The aneurysm and the necrotic tissue were resected, the native aortic valve was oversewn and a new left ventricular outflow tract was created from the apex of the ventricle to the ascending aorta. The infant is currently well and without residual defects.

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